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Case Report

Angiography and En Face Optical Coherence Tomography Findings in Acute Syphilitic Posterior Placoid Chorioretinopathy

Aina Moll-Udina Juan P. Figueroa-Vercellino Victor Llorenç
Lucía Miguel Alfredo Adán

Uveitis Unit, Hospital Clinic de Barcelona, Instituto de Oftalmología, Facultad de Medicina, Universidad de Barcelona, Barcelona, Spain

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Acute syphilitic posterior placoid chorioretinopathy · Optical coherence tomography angiography · Optical coherence tomography en face · Ocular syphilitic manifestation · Uveitis · Syphilis

Abstract
Acute syphilitic posterior placoid chorioretinopathy (ASPPC) is one of the rarest ocular manifestations of syphilis. The pathophysiology of this entity is still unknown. We report the outer retinal findings on en face optical coherence tomography (OCT) and the alteration of choriocapillaris flow findings on OCT angiography in a patient with ASPPC at the time of presentation, after penicillin treatment completion and during follow-up.

Introduction

Syphilis is a systemic infectious disease caused by the spirochete *Treponema pallidum* often known as the great masquerader because of the variety of ocular signs it can display. Acute syphilitic posterior placoid chorioretinopathy (ASPPC) is a rare ocular manifestation of

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Aina Moll-Udina, MD
Hospital Clinic de Barcelona, Instituto de Oftalmología
1 Sabino de Arana
ES–08028 Barcelona (Spain)
E-Mail moll@clinic.cat

syphilis, first described by Gass et al. [1] in 1990 as a large, yellowish, circular or oval placoid lesion at the level of the retinal pigment epithelium (RPE) in or near the macula. The pathophysiology of ASPPC is not completely understood; it has been postulated to be an inflammatory reaction or immune complex deposition at the level of the choroid-RPE-photoreceptor complex [1]. We describe a case with multimodal imaging, allowing a better understanding of the pathophysiology in a patient diagnosed with ASPPC, supporting that the choriocapillaris may be primarily involved.

Case Report

A 46-year-old man presented with 1-month blurry vision in the right eye (OD) and no other systemic symptoms. He suffered from temporal lobe epilepsy due to a benign astrocytoma, operated in 1993 and controlled with antiepileptic drugs. Visual acuity (VA) was counting fingers in his OD and 20/20 in his left eye (OS). Anterior examination was unremarkable. Funduscopic examination revealed a yellowish placoid lesion occupying the whole posterior pole in his OD, and there was an apparently normal funduscopic examination in his OS (Fig. 1). Ultra-wide-field fundus autofluorescence demonstrated hyperautofluorescence corresponding to the placoid lesion in OD and an area of hyperautofluorescence superotemporal to the optic disk in OS (Fig. 1). Spectral-domain optical coherence tomography (SD-OCT; Cirrus HD OCT®, Carl Zeiss, USA) B-scan showed a disruption of the ellipsoid zone and external limiting membrane, hyperreflective granular RPE changes, and areas of punctate hyperreflectivity in the underlying choroid in the OD (Fig. 1e). En face OCT revealed multiple hyperreflective clumps in the outer retina in the OD (Fig. 2), and OCT angiography (OCT-A; Angioplex®) showed areas of flow void at the level of the choriocapillaris in both eyes (Fig. 3). Blood analysis revealed reactivity to the venereal disease research laboratory (VDRL) (>1/1,024) and positivity antibodies against membrane protein A of *T. pallidum* by ELISA (IgG and IgM), indicating acute syphilitic infection. Lumbar puncture was negative for VDRL and for IgM anti-*T. pallidum* but positive for IgG, and pleocytosis was seen in the cerebrospinal fluid.

Brain magnetic resonance imaging was unremarkable. Serum anti-HIV antibodies were negative. The patient underwent the standard treatment for neurosyphilis, i.e., intravenous penicillin G at a dosage of 24 million units per day for 14 days and i.v. pulse of methylprednisolone. After he completed the antibiotic treatment, his VA improved to 20/20 in both eyes, and although the outer retinal lesions were completely restored, choriocapillaris reperfusion remained incomplete at 3 months of follow-up (Fig. 3).

Discussion

The most common presentation of syphilis in the eye is uveitis [2, 3] as chorioretinitis. ASPPC defines a variant of syphilitic chorioretinitis [1]. The first reported cases were patients coinfected with HIV [1], but nowadays, it has been reported in immunocompetent patients [3–5] as in the present case. In fact, Fonollosa et al. [3] did not observe any patient coinfected with HIV in the 8 patients with ASPPC reported from Northern Spain. In contrast, Pichi et al. [6] observed a rate of 31.5% of HIV coinfection, and a rate of 40% was observed by Eandi et al. [7].

Pichi et al. [6] described localized hyperautofluorescence in the area of the placoid lesion as well as focal intense hyperautofluorescence in some eyes, as in our patient. These findings

seem to be due to subretinal deposition of RPE-photoreceptor complex material and an incomplete phagocytosis of outer segments [7].

Brito et al. [8] described an acute disruption of the external limiting membrane and ellipsoid layer, the same findings as in our patient. Outer retina disruption was completely reversible after penicillin treatment.

En face OCT allowed us to observe a photoreceptor alteration seen as multiple hyperreflective clumps in the ellipsoid/RPE layer. Lima et al. [9] support the assumption that these dots corresponded to the hyperreflective nodularity of the RPE seen on SD-OCT, and Zett et al. [4] could co-localize exactly these lesions to the hyperfluorescent dot-like lesions seen on fundus autofluorescence imaging.

Similar to previous reports [4, 8–10], nodular elevations of the RPE were seen on B-scan OCT, which are typical in this entity and were observed in our patient as well. These RPE changes seem to be well correlated with hyperautofluorescent dots seen on fundus autofluorescence imaging [11]. The hyperreflective pinpoint lesions seen on SD-OCT in the choroid have been described in the literature [6, 8] and may represent an inflammatory reaction in the choroid and choriocapillaris or an infected infiltrated choroid.

The pathophysiology of ASPPC is not clearly understood, although Gass et al. [1] postulated an inflammatory reaction or an immune complex dysfunction at the level of the choroid-RPE-photoreceptor complex resulting in a placoid lesion and photoreceptor dysfunction. Some authors [1, 6] proposed a local choroidal hypoperfusion and/or blockage of the choroidal fluorescence by the overlaying infected RPE as seen in indocyanine green angiography.

We support the assumption that the inflammatory origin could be in the choriocapillaris due to our multimodal imaging findings. A large area of nonperfusion in the clinically affected eye (OD) was observed at the level of the choriocapillaris on OCT-A and also a slow nonperfusion area in OS, in which VA was 20/20. Photoreceptor and RPE disturbances might be a consequence of a reduced choriocapillaris flow.

While bilateral choriocapillaritis with hypoperfusion was observed in both eyes, a really different affection was observed both on SD-OCT and en face OCT, the OD being much more affected. These structural differences support the inflammatory origin because a primary choriocapillaritis was present in both eyes although showing almost no external retinal modifications in the OS. Furthermore, after the treatment, the outer retina had a complete recovery, while the choriocapillaris just showed a little perfusion improvement. On the one hand, this might be due to the initial severity of the hypoperfusion. Tsui et al. [10] described small and focal areas of hypoperfusion which were reversed promptly after 2 months of follow-up. In our case, choriocapillaris flow void was more severe in the initial OCT-A, which might have led to a slower recovery of the choriocapillaris perfusion. Even in the 3 months of follow-up, hypoperfusion still existed in both eyes.

On the other hand, this lack of correlation might be due to an initial ischemic choroidal infarction found in our patient. The initial choriocapillaritis generated an inflammation in the outer retina, which promptly recovered after treatment, as seen on the OCT B-scan, whereas the choroidal infarction might take much longer to recover.

To our knowledge, this is the first ASPPC patient in which there was a clear area of flow void in the choriocapillaris layer seen on OCT-A, as it has been demonstrated in acute posterior multifocal placoid pigment epitheliopathy [12, 13].

ASPPC can alert us of an acute syphilitic infection and, for the ophthalmologist, plays a vital role in the detection and treatment of this infection. If syphilis is promptly recognized and properly treated, it has an excellent functional prognosis. Angiography and en face OCT segmentations can be useful for the differential diagnosis and follow-up of this rare disease.

Statement of Ethics

The patient has consented to the submission of the case report.

Disclosure Statement

The authors declare no commercial or proprietary conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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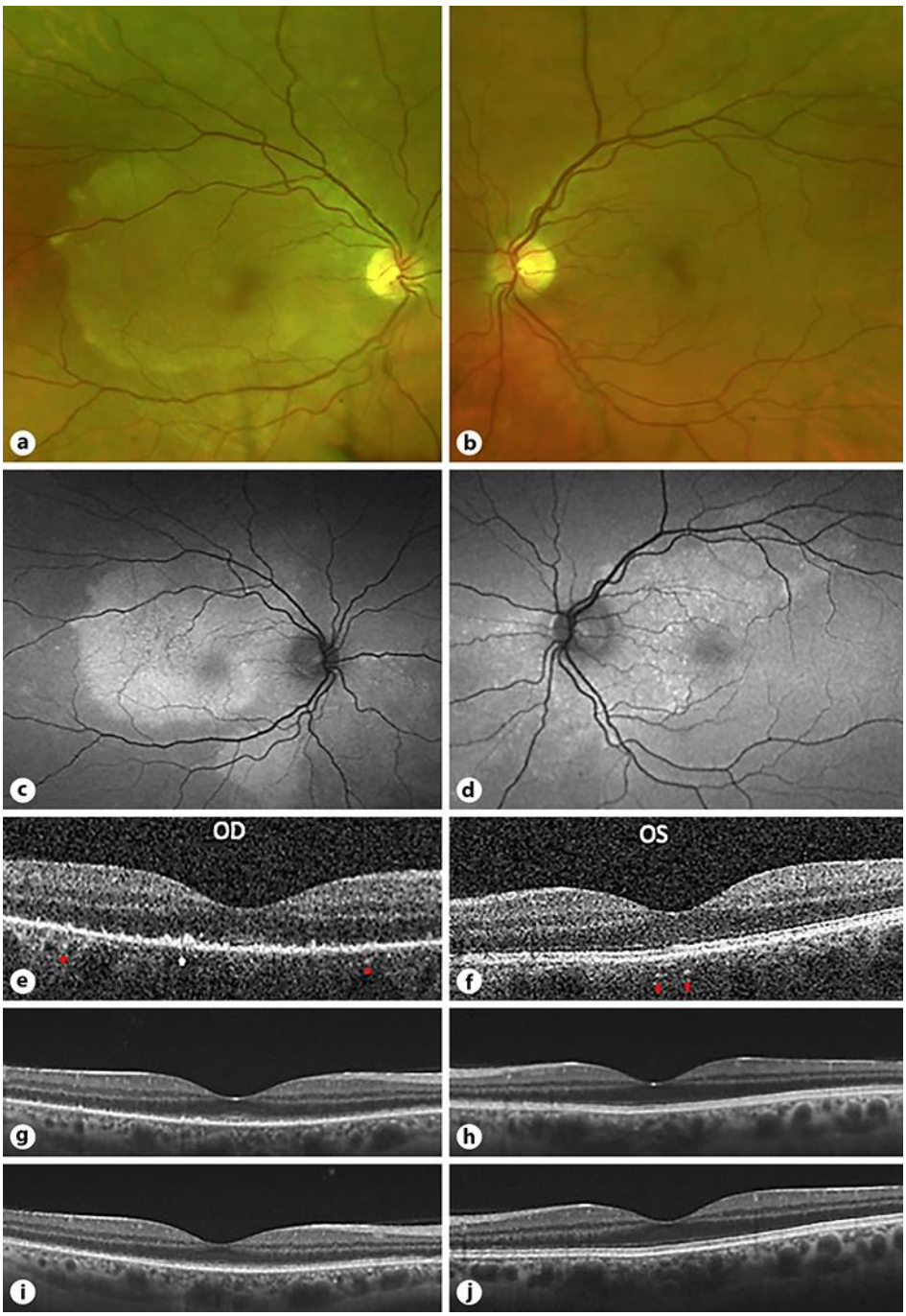

Fig. 1. Color fundus photograph and autofluorescence of the right eye (**a**, **c**) and the left eye (**b**, **d**). **a** Yellowish placoid lesion occupying the whole posterior pole. **b** Apparently normal fundus. **c** Hyperautofluorescent lesion corresponding to the placoid lesion. **d** Hyperautofluorescent area occupying half of the macula not appreciated in **b**. **e** SD-OCT B-scans reveal a disruption and loss of the ellipsoid zone (EZ) and external membrane limiting (ELM) layers, nodular elevations of the retinal pigment epithelium (RPE) (white arrow), and hyperreflective pinpoint lesions inside the choroid (red arrows) in the right eye. **g** After 7 days of treatment, swept-source OCT B-scan shows a decrease in nodular elevations. **i** At 14 days of penicillin treatment, a nearly total remodellation of the EZ and ELM layer and a complete resolution in both RPE nodulations and punctate hyperreflectivity in the choroid are demonstrated. SD-OCT of the left eye (**f**) shows a discontinued ELM layer but a pretty much conserved EZ and a few hyperreflective lesions inside the choroid (red arrows) that had completely recovered after the treatment (**h**, **j**).

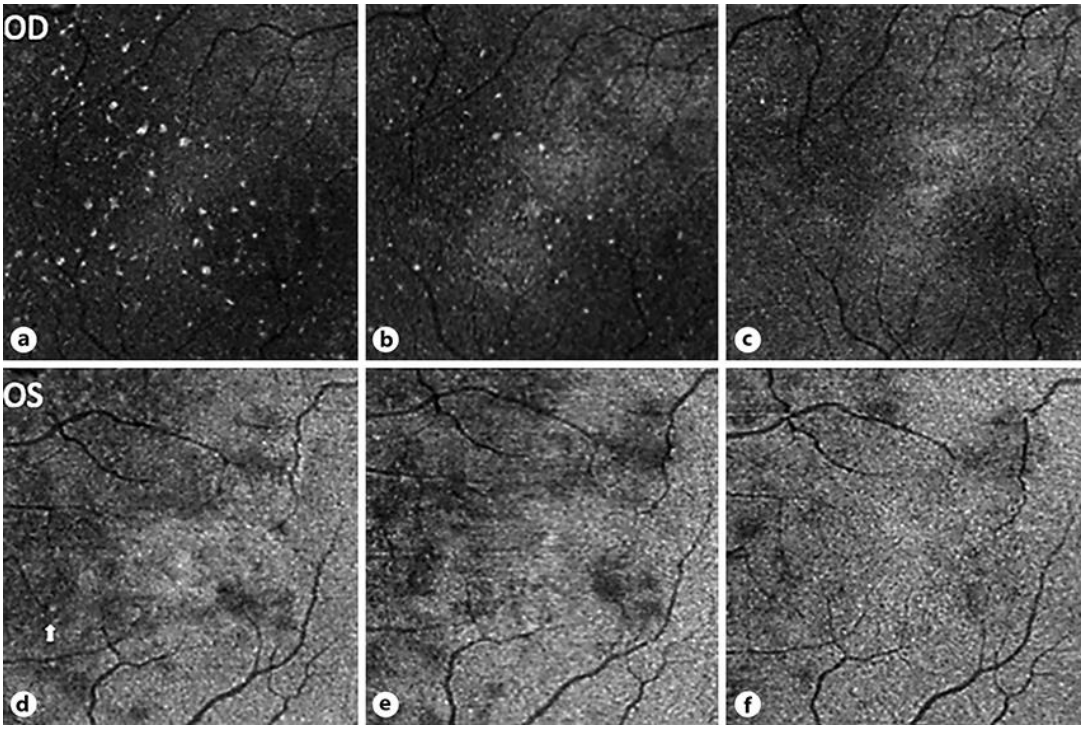

Fig. 2. In the top row, 3 × 3 mm en face OCT at the level of the ellipsoid layer shows several hyperreflective dots above a hyporeflective background in the right eye (**a**), a decrease of these lesions after 1 week of penicillin treatment (**b**), and the disappearance of them after completed treatment of 14 days (**c**). In the bottom row, 3 × 3 mm en face OCT at the level of the ellipsoid layer reveals a hyporeflective area corresponding to the placoid lesions on fundus autofluorescence (Fig. 1d) with a few hyperreflective dots (**d**, white arrow) in the left eye. The lesion had nearly recovered after 14 days of penicillin treatment (**f**).

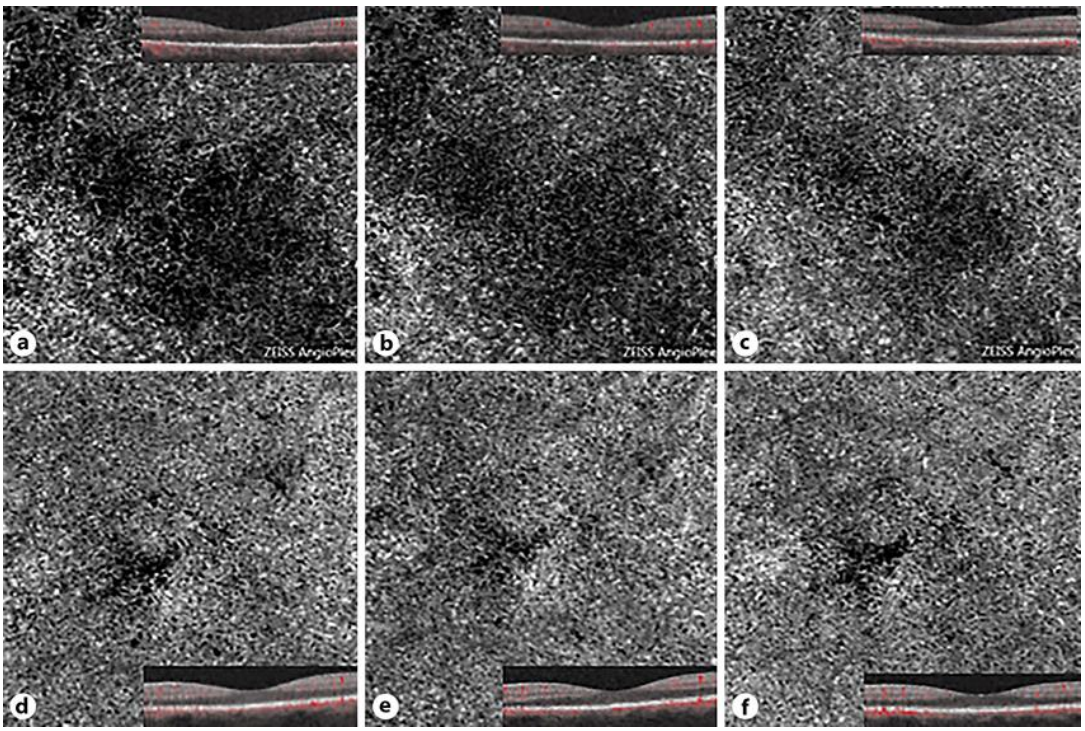

Fig. 3. 3 × 3 mm OCT-A of choriocapillaris layer of the right eye (**a–c**) and the left eye (**d–f**). Large area of nonperfusion before i.v. penicillin treatment (**a**) that recovered slightly after 1 week of treatment (**b**) and after 3 months of follow-up (**c**). Small area of nonperfusion before penicillin treatment (**d**) that remains stable after 1 week (**e**) and at 3 months of follow-up (**f**). Each OCT-A image has its correlation with the OCT B-scan superimposed flow signal.